

## Case Report

# Unusual Presentation of Abdominal Castleman's Disease

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Castleman's disease is a rare lymphoproliferative disorder of uncertain origin. We report an unusual presentation of Castleman's disease in the transverse mesocolon that mimicked a vascular gastrointestinal stromal tumour and review the literature surrounding this peculiar entity. [*Asian J Surg* 2006;29(3):153–6]

**Key Words:** Castleman's disease, lymphoproliferative, mesocolon

## Introduction

Castleman's disease is a rare lymphoproliferative disorder of uncertain origin. We report an unusual presentation of Castleman's disease in the transverse mesocolon that mimicked a vascular gastrointestinal stromal tumour and review the literature surrounding this peculiar entity.

## Case report

A 28-year-old man presented with vague abdominal discomfort and mechanical low back pain for about 2 months. Physical examination was unremarkable, laboratory data were within the normal limits and chest X-ray was normal. Lumbosacral X-rays (Figure 1) revealed no bony abnormalities but showed an amorphous opacity between the transverse processes of L3 and L4 on the right suggestive of retroperitoneal calcification. Computed tomography (CT) scan (Figure 2) showed a well-defined vascular soft tissue mass measuring 4 × 3.5 cm situated inferior to the second part of the duodenum and adjacent to the inferior vena cava. It bore a focus of calcification that was surrounded by soft tissue of low density representative of necrosis. Oesophagogastrroduodenoscopy merely showed a subtle eccentric deformity at the junction of the second and third parts of the duodenum causing

extrinsic compression. With clinical features suspicious of a duodenal gastrointestinal stromal tumour, surgical excision was planned in view of its size and the patient's symptoms.

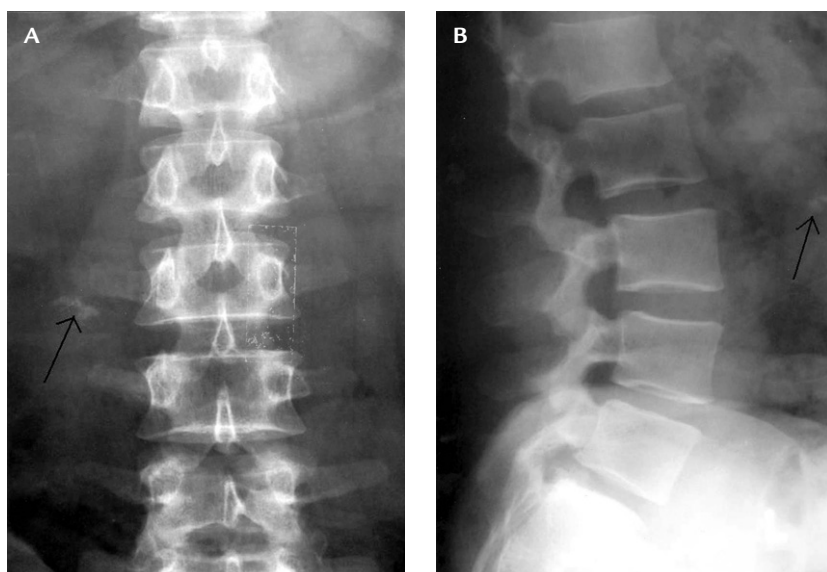
Intraoperatively, the tumour was found within the transverse mesocolon and measured 5.5 × 3.5 × 3.5 cm. It had a fleshy consistency suggestive of a lymphomatous lymph node (Figure 3). A thorough exploration revealed no intra-abdominal lymphadenopathy or any visceral abnormalities. The tumour was closely related to the superior mesenteric artery (SMA) on its medial aspect with multiple feeding vessels. It was meticulously dissected with a gross margin of 1 cm. Feeding vessels were ligated and divided without injuring the SMA or compromising bowel viability. Histological examination demonstrated typical features of hyaline vascular type of Castleman's disease (Figure 4). During the 1-year follow-up, the patient was well with no evidence of recurrent disease.

## Discussion

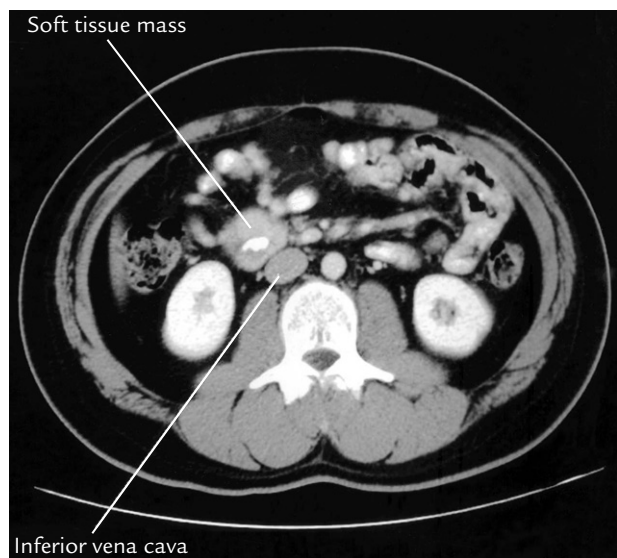
Castleman's disease was first described in 1954 in 13 patients with giant mediastinal lymph nodes resembling thymoma.<sup>1</sup> Since then, it has been referred to as angiofollicular lymph node hyperplasia, giant lymph node hyperplasia, lymphoid hamartoma, angiomatous lymphoid hamartoma,

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**Figure 1.** Lumbosacral X-rays show an amorphous opacity (arrow) suggestive of retroperitoneal calcification.

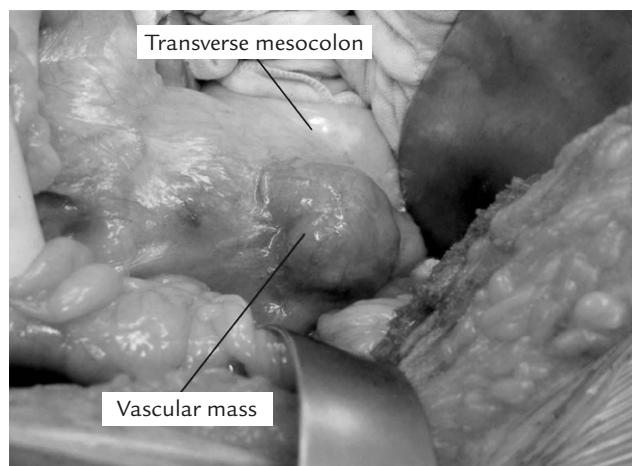


**Figure 2.** Computed tomography show the close proximity of the soft tissue mass (with a calcified centre) to the inferior vena cava.

benign lymphoma or follicular lymphoreticuloma, only to account for its uncertain aetiology. Theories including chronic low grade inflammation, hamartomatous process, an immunodeficient state and autoimmune process have been proposed as likely pathogenetic mechanisms.<sup>2,3</sup>

Based on histological characteristics, Keller et al classified this disease into two variants: the hyaline vascular type (HV) and the plasma cell (PC) type.<sup>3</sup>

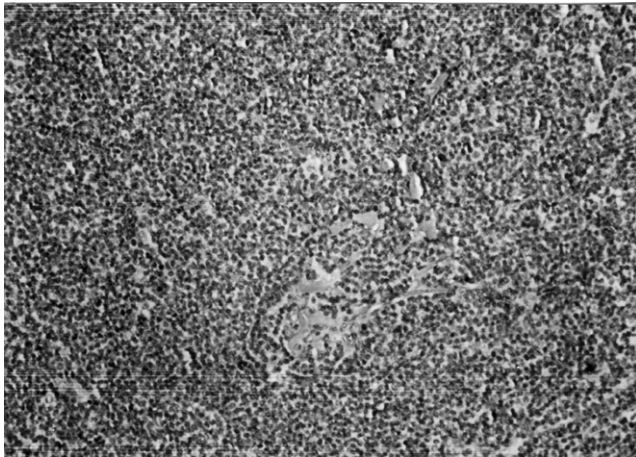
The HV variant consists of small distinct vascular follicles surrounded by palisading layers of small lymphocytes with radially arranged capillaries penetrating the germinal centres while the PC variant demonstrates larger lymphoid follicles and a markedly less vascular interfollicular



**Figure 3.** Intraoperative findings of the vascular mass with multiple feeding vessels within the transverse mesocolon.

stroma where PCs predominate. The HV variant is by far more common, comprising 91% of reported cases and largely runs a benign course, resulting in symptoms in only 3% of cases,<sup>4</sup> which are largely discovered incidentally, as in our case. In contrast, though less common, the PC variant tends to be more clinically aggressive with systemic manifestations such as fever, anaemia, hypergammaglobulinaemia and an increase in acute phase proteins in up to 50% of cases. Interleukin-6 (IL-6) is reported to play a key role in these systemic features,<sup>2</sup> with clinically documented decrease in its levels and disappearance of symptoms following surgical removal.

Though first reported occurring in the mediastinum,<sup>1</sup> virtually all lymph node-bearing regions and even non-nodal tissues may be involved.<sup>2,3</sup> Its clinical presentations



**Figure 4.** Blood vessels with hyalinized walls traverse the centre of a germinal centre, a typical feature of hyaline vascular Castleman's disease.

can be either localized or multicentric. Localized disease is usually asymptomatic and involves the mediastinum in 70% of cases. Isolated cases have been reported in the central nervous system, orbit, neck, axilla, skeletal muscles, the retroperitoneum, pancreas and the mesentery.<sup>2-4</sup> In particular, mesenteric disease is rare with only 22 cases reported in the English literature to date. Of interest, localized disease is predominantly of the HV type and potentially curable by surgical resection. Though surgical removal might not always be necessary for asymptomatic Castleman's disease, operative management is frequently unavoidable since preoperative diagnosis is evasive.

In contrast, multicentric disease is clinically more aggressive with systemic manifestations and potentially fatal infective complications. It has a tendency to involve the peripheral lymph nodes and is predominantly of the PC variant. It may run one of the four courses: relapse and remission, stable and persistent, rapidly fatal or transformation into malignant lymphoma. Systemic therapy in the form of steroids and chemotherapeutic agents such as cyclophosphamide, doxorubicin and vincristine have been tried with varying success rates, while administration of murine anti-IL-6 antibody has been reported to produce transient improvement in symptoms and laboratory values.<sup>5</sup>

Though definitive diagnosis necessitates histological analysis, radiological features may be helpful. HV lesions often demonstrate fine calcification that is evident even on plain X-rays, as in our case. On CT or magnetic resonance imaging, lymphoid lesions, which are involved, typically demonstrate homogeneous contrast enhancement. This distinguishes Castleman's disease from other masses

such as thymomas or lymphomas, which generally show no enhancement on CT scan.<sup>6</sup> Being highly vascular lesions, these masses characteristically show a prolonged dense tumour "blush" in the capillary phase of angiography. This can be used to delineate the hypertrophied feeding vessels that supply these lesions. Preoperative embolization has been advocated to facilitate surgical excision and decrease intraoperative bleeding, but this is not widely practiced.<sup>7</sup>

Complete surgical excision is the mainstay of treatment and is virtually curative in all cases reported so far, with a 5-year survival rate approaching 100%.<sup>3,4</sup> However, local recurrence has been reported after subtotal or partial resection. Radiation therapy has been used with varying success in unresectable lesions.<sup>8</sup>

The appearance of associated malignant tumours has been occasionally reported and may be the result of a slow hyperplastic process, appearing as lymphomas in the PC type<sup>9</sup> and vascular tumours resembling Kaposi's sarcoma in the HV type.<sup>10</sup> These have been reported to develop as long as 8 years after the initial diagnosis, hence, lifelong follow-up is recommended. As Castleman's disease remains a rare and poorly understood condition, there is no existing best way to follow-up these patients. For cases of localized HV type of Castleman's disease similar to ours, we propose 6–12 monthly physical examinations after surgical excision with abdominal CT scan or ultrasound evaluation every 3–5 years or whenever symptoms arise.

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